Management of Thalassemia at Sultan Qaboos University Hospital, Muscat, Oman 1991 -2016

1st International Working Group on Thalassemia:
IS IT TIME TO REVISIT CLASSIFICATION OF THALASSEMIA SYNDROMES ?

Prof Shahina Daar

Following the Policy of the National Regulation 3.3 , page 17, on CME disclosures, dated 5 November 2009, and on behalf of the Provider , - Collage S.p.A.- n. 309

I, SHAHINA DAAR HERE DECLARE

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I have no relevant personal financial relationship in the medical/health field.

DISCLOSURE OF PROMOTIONAL TALKS

I have not presented any promotional talks for any pharmaceutical companies within the past 12 months

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15-16 September, 2017 *Shahina Daar*

- Transfusion & Chelation
- MRI T2*
- Survival

Transfusion

Sultan Qaboos University Hospital Thalassemia Unit opened in 1991 and almost all patients with TM were transferred over the next 12 months from other hospitals.

Health care services in Oman initiated in 1971 with inadequate transfusion. Therefore

- Many patients had been splenectomised
- Many had short stature and thalassemic facies

Current policy

- Maintain Hb >10gm/dl for children and >9 gm/dl for adults
- Leucoreduction either pre storage or at bedside
- Routine pre transfusion antibody screening
- Extended phenotype matching for all TI patients

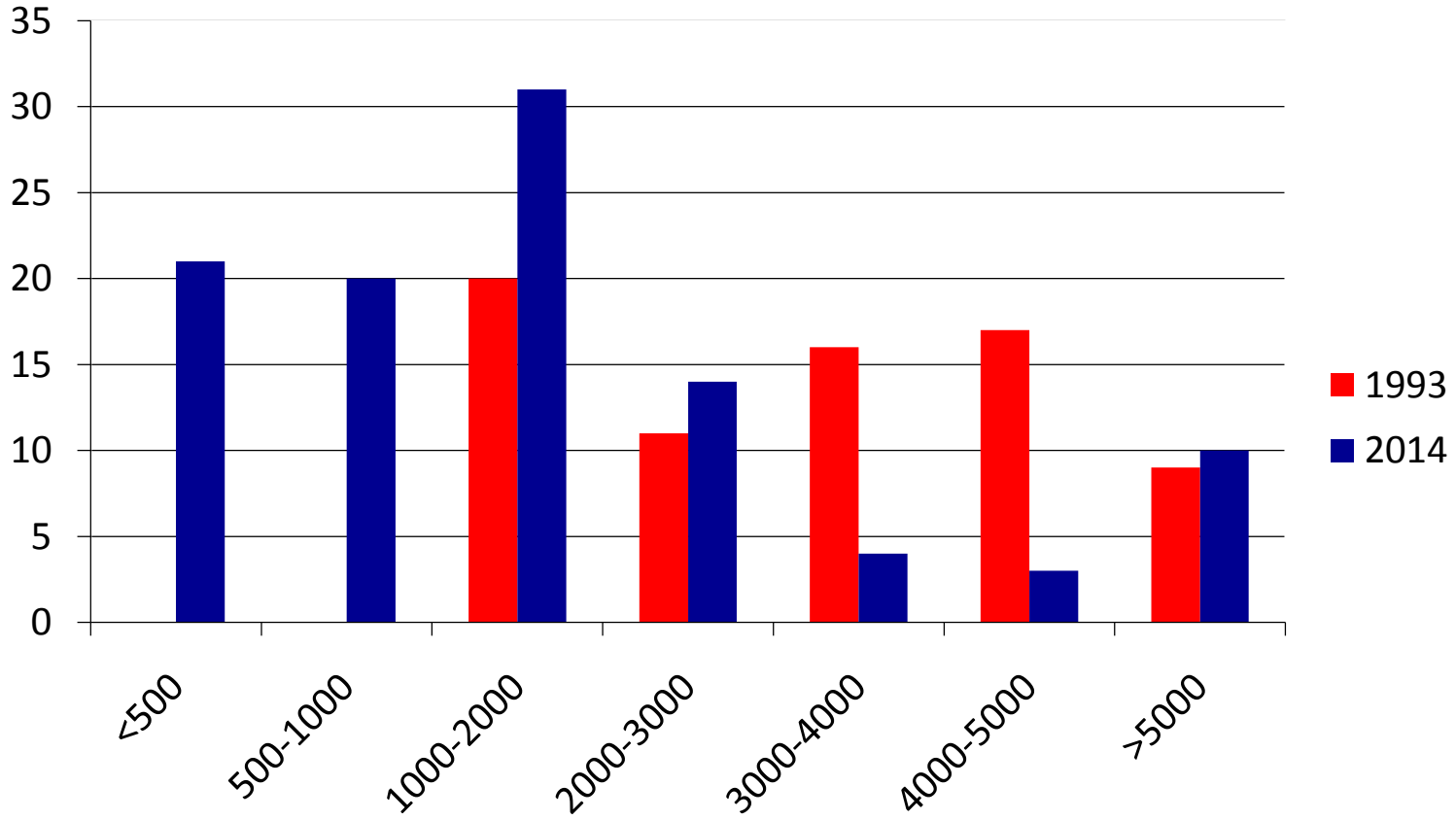
Recommendations

- Extended phenotype at diagnosis before any transfusion
- Add C & E matching when stocks allow

CHELATION:

- At time of transfer, **only 4 of 91 patients were on regular chelation** - the remainder had ONLY received DFO at time of transfusion.
- By 1992, all patients who agreed were on regular DFO
- 2000 -2001 most patients were on sequential DFO + DFP
- 2005 – ESCALATOR TRIAL
- 2009 – DFX available
- 2012 - DFX + DFO/DFP
- 2014 - 24hour IV DFO Baxter pump, limited numbers

S FERRITIN 1993 /2014



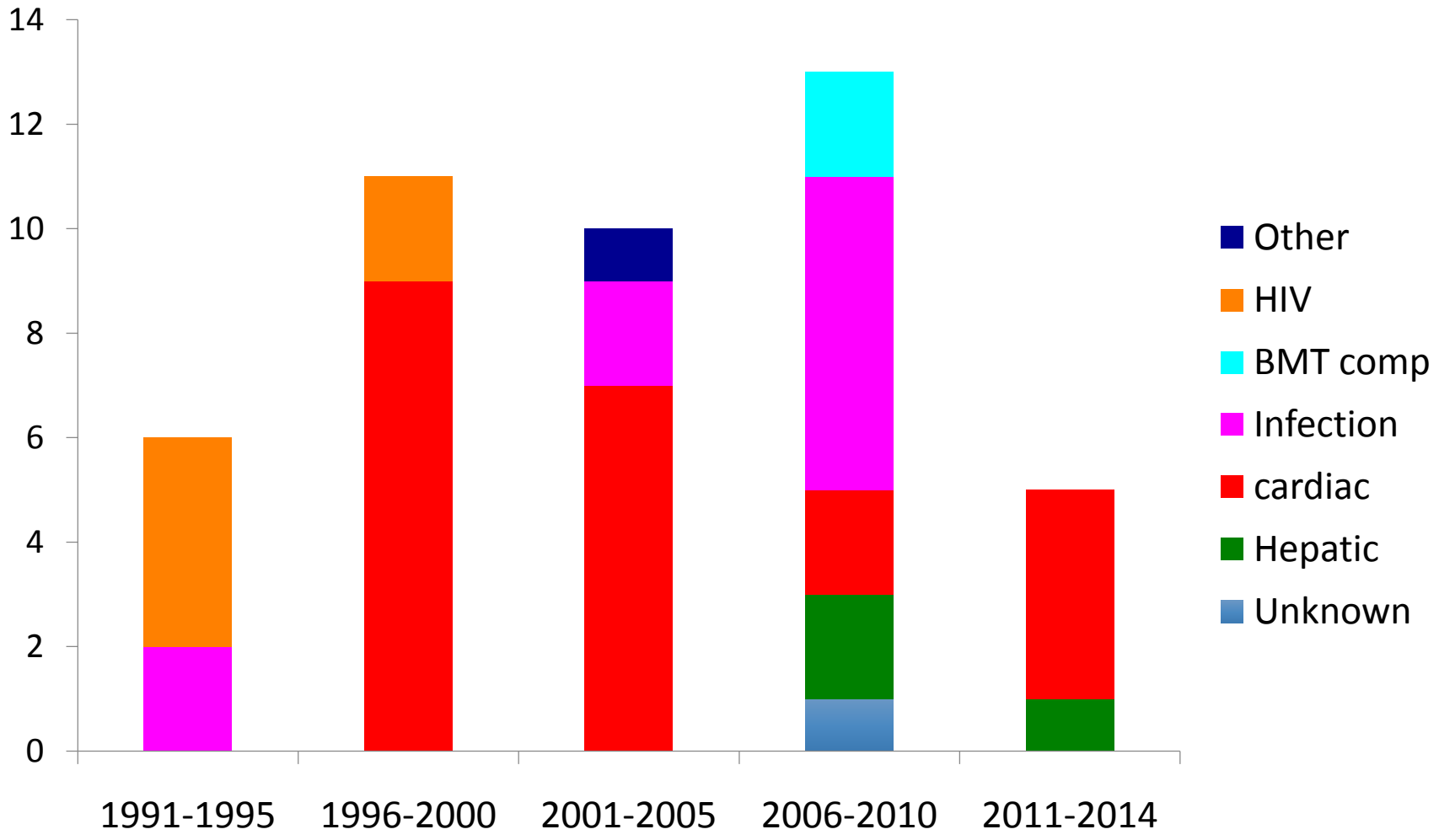
MRI Cardiac T2* - Started September 2006 : OUTCOME

Cardiac T2*(ms)	Total at Baseline 2006	Died	< 10 2016	10-19.9 2016	>20 2016	2016
< 10	24	9	2	3	8	5
10-19.9	30	4	3	9	16	15
>20	52	6	0	3	43	67
Total	106	19				

MRI Liver T2* - Started September 2006 : OUTCOME

Liver mg/gdw	2006	Died	>15 2016	7-15 2016	3-6.9 2016	<3 2016	2016
>15	36	9	13	7	5	2	21
7-15	33	5	6	7	7	8	19
3-6.9	24	1	2	5	8	8	23
<3	13	5	0	0	3	5	23
Total	106	20					

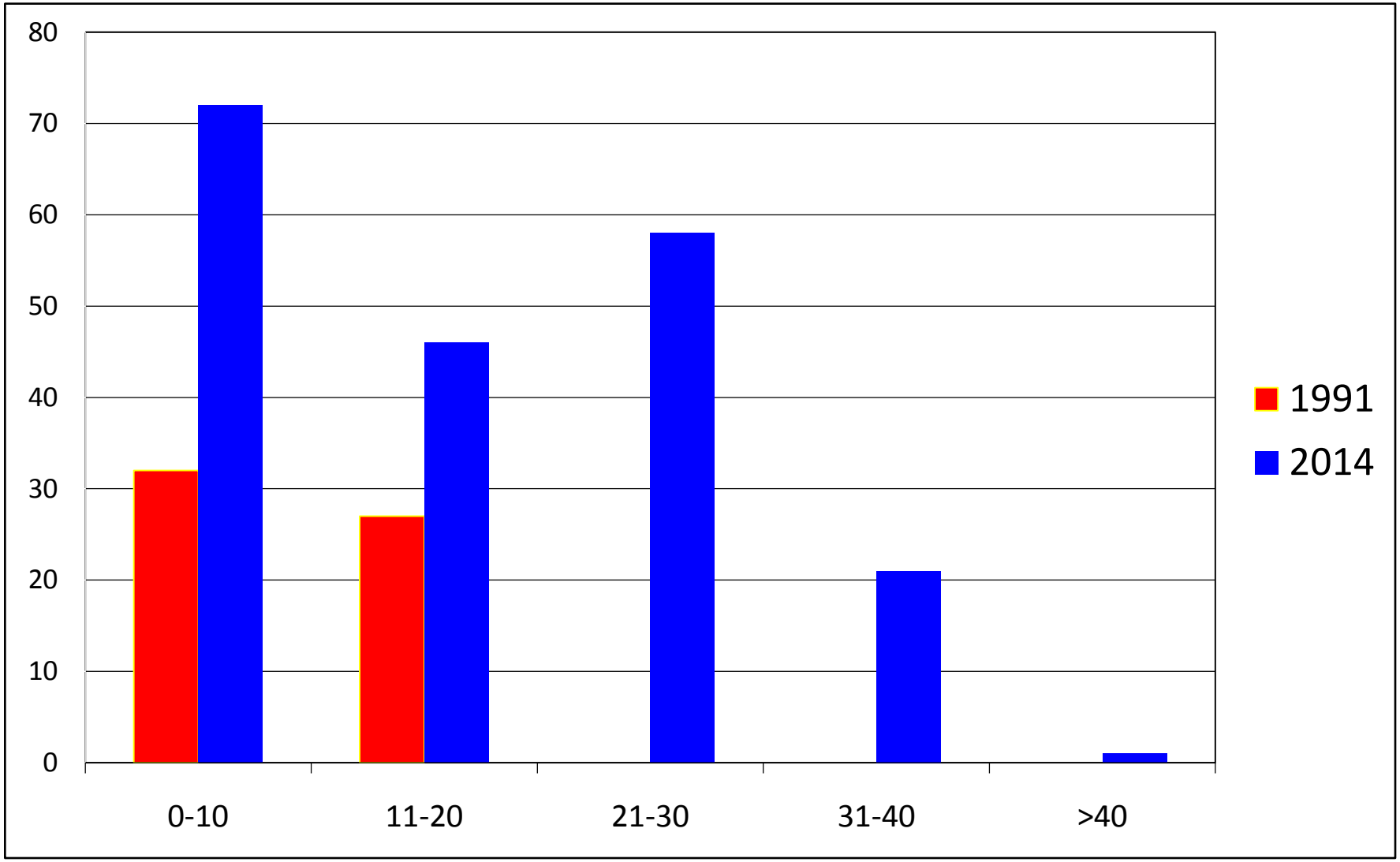
Change Causes of Death in Unit since 1991



Hepatitis

- Government policy to vaccinate all children for HBV
- Thalassemia patients get screened for HBV, HCV, HIV every 6 months
- High percentage of our pts who were previously infected with HCV cleared virus spontaneously
- 15 of our patients have seroconverted to HCV despite ELISA testing
- We still do not have NAT testing for blood units, although plans are there
- 2 patients have died with HCC, one of whom got HCC 8 yrs after successful Rx and LIC of 1.1mg/gdw
- Harvoni available free and all our thalassemia patients have been successfully treated

AGE DISTRIBUTION OF TM AT SQUH (BOTH ADULT AND CHILDRENS UNIT)



Thank You